

Computed Tomography / Tomodensitométrie

Imaging Appearances of Congenital Thoracic Lesions Presenting in Adulthood

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Abstract

Many congenital lesions of the thorax are detected for the first time in adulthood when they can simulate a wide range of pathologies, including infection and neoplasia. They can be broadly classified into tracheobronchial, parenchymal, vascular, and combined parenchymal/vascular abnormalities. An awareness of their typical imaging features enables a confident diagnosis and helps direct appropriate patient management.

Abrégé

Bon nombre de lésions congénitales du thorax sont décelées pour la première fois à l'âge adulte, alors qu'elles peuvent simuler toutes sortes de pathologies, notamment des infections et des néoplasies. On peut en gros les considérer comme des anomalies trachéobronchiques, parenchymateuses, vasculaires ou parenchymateuses-vasculaires combinées. Une bonne connaissance de leurs caractéristiques d'imagerie type permet de poser un diagnostic fiable et favorise une prise en charge adéquate du patient.

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Key Words: Congenital; Thoracic; Tracheal bronchus; Tracheobronchomegaly; Cystic adenoid malformation; Bronchogenic cyst; Pulmonary hypoplasia; Scimitar syndrome; Pulmonary sequestration

Many congenital lesions of the thorax are detected for the first time in adulthood when they can simulate a wide range of pathologies, including infection and neoplasia. Although some of these conditions are simply incidental findings that require no specific management, others potentially can be life threatening. This review illustrates the plain film and cross-sectional imaging appearances of these conditions, which can be broadly classified into tracheobronchial, parenchymal, vascular, and combined parenchymal/vascular abnormalities (Table 1).

Abnormalities of the Tracheobronchial Tree

Tracheal Bronchus

Tracheal bronchus is an anatomical variant whereby an anomalous airway arises from the lateral wall of the trachea. The majority of tracheal bronchi are right sided and arise within 2 cm of the carina; however, left tracheal bronchi have also been described. Tracheal bronchus has a prevalence of 0.1%–2% from bronchoscopic studies [1] and is more common in patients with Down syndrome. There are many possible anatomical configurations, including a displaced right upper-lobe bronchus, giving rise to all 3 upper-lobe bronchopulmonary segments, or, more commonly, it may just supply the right apical segment [2].

Tracheal bronchus is most often an incidental finding, although, in the critical care setting, attention is drawn to its possibility when persisting lobar atelectasis is noted in

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Table 1

Developmental thoracic lesions that may present in adulthood

Abnormalities of the tracheobronchial tree
Tracheal bronchus
Tracheobronchomegaly
Bronchial atresia
Bronchogenic cyst
Abnormalities of the lung parenchyma
Pulmonary agenesis/aplasia
Pulmonary hypoplasia
CAM
Abnormalities of the vasculature
Vascular rings
Arteriovenous malformation
PAPVR
Combined parenchymal/vascular abnormalities
Scimitar syndrome
Pulmonary sequestration

spite of an adequately positioned endotracheal tube (Figure 1). It may also be associated with recurrent pneumonia, chronic bronchitis, and bronchiectasis. Computed tomography (CT) can eloquently demonstrate the aberrant bronchus and its precise anatomical configuration (Figure 2). Surgical resection is considered in severely symptomatic cases [3].

Tracheobronchomegaly

Tracheobronchomegaly (Mounier-Kuhn syndrome) is a rare condition characterized by atrophy of elastic and smooth muscle fibers in the trachea and central bronchi, which become markedly dilated. An abrupt change to normal

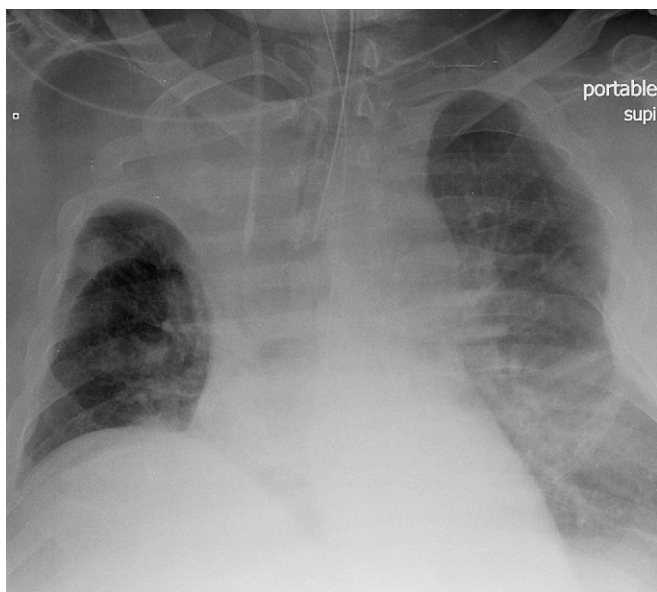


Figure 1. Chest radiograph of a 52-year-old man admitted to intensive care after a road traffic accident. An endotracheal tube is appropriately positioned, but there is right upper-lobe collapse. These changes persisted, and a subsequent CT scan showed a right tracheal bronchus as the cause.



Figure 2. Coronal reformatted CT image, showing a right-sided tracheal bronchus.

calibre occurs at the fourth or fifth order bronchial divisions [4]. It has been reported as a familial trait with an autosomal recessive pattern of inheritance [5] and also in association with Ehlers-Danlos and Marfan syndrome; however, in many cases, it is thought to be an acquired condition. Inhalation of chemical irritants and barotrauma from prolonged mechanical ventilation are recognized associations.

Most cases present in adult life with chronic cough, excessive sputum production, and recurrent chest infections as a consequence of impaired clearance of secretions [6]. Diagnostic criteria based on radiologic findings are tracheal and main-stem bronchial diameters that exceed more than 3 standard deviations from the mean. A tracheal diameter larger than 26 mm in men and 23 mm in women is considered diagnostic. CT enables a more comprehensive assessment of the central airways and may show supportive features, such as tracheobronchial diverticulosis, cystic bronchiectasis, and complete collapse of the central airways on expiration (Figure 3).

Bronchial Atresia

Bronchial atresia is a rare condition caused by congenital occlusion of a proximal bronchus, most likely the consequence of a vascular insult in early fetal life [7]. The bronchus immediately distal to the occlusion fills with mucoid secretions, but the distal lung parenchyma develops normally and becomes hyperaerated via collateral air-drift pathways. The left upper-lobe bronchus is most frequently involved [8]. Most cases are incidental findings on chest radiographs;

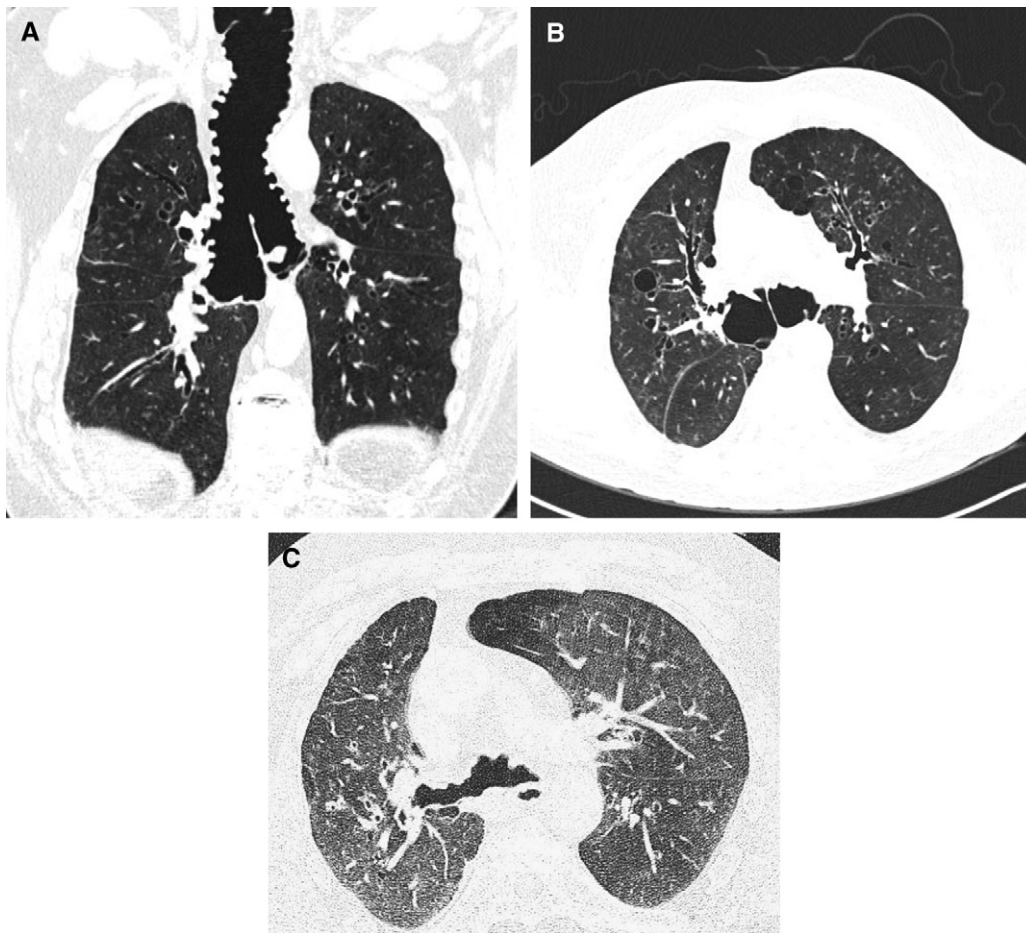


Figure 3. A 45-year-old man with tracheobronchomegaly who presented with recurrent chest infections. (A) Coronal reformatted CT image, showing marked dilatation of the trachea and main bronchi, which have a corrugated appearance. (B) Transaxial CT image, showing dilatation of both main bronchi and cystic bronchiectasis. (C) Expiratory phase transaxial CT image, demonstrating collapse of the central airways and associated air trapping.

however, patients may present with a history of recurrent chest infections. Surgical resection of the involved segment is curative in such cases [9].

Chest radiograph characteristically shows a well-defined central ovoid or linear branching mass with distal

hyperexpansion (Figure 4A). CT shows a dilated mucus-filled bronchus with peripheral emphysematous lung (Figure 4B). Expiratory phase imaging highlights the air-trapping, and multiplanar reformats are useful in distinguishing mucoid impaction from an endobronchial tumour [10].

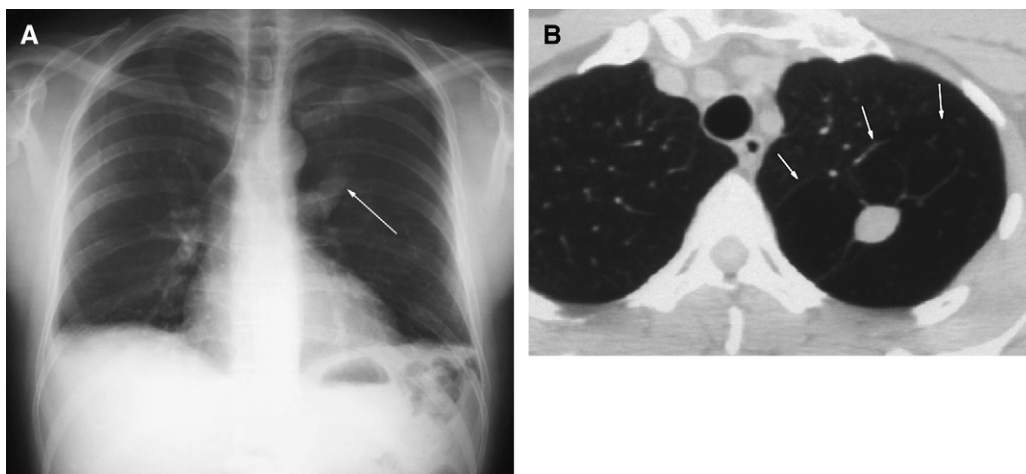


Figure 4. An asymptomatic 34-year-old man with no chest symptoms. (A) Chest radiograph, showing left upper-lobe hyperexpansion and a central ovoid mass (arrow). (B) Transaxial CT image, showing a dilated mucus-filled bronchus and peripheral emphysematous lung (delineated by arrows).

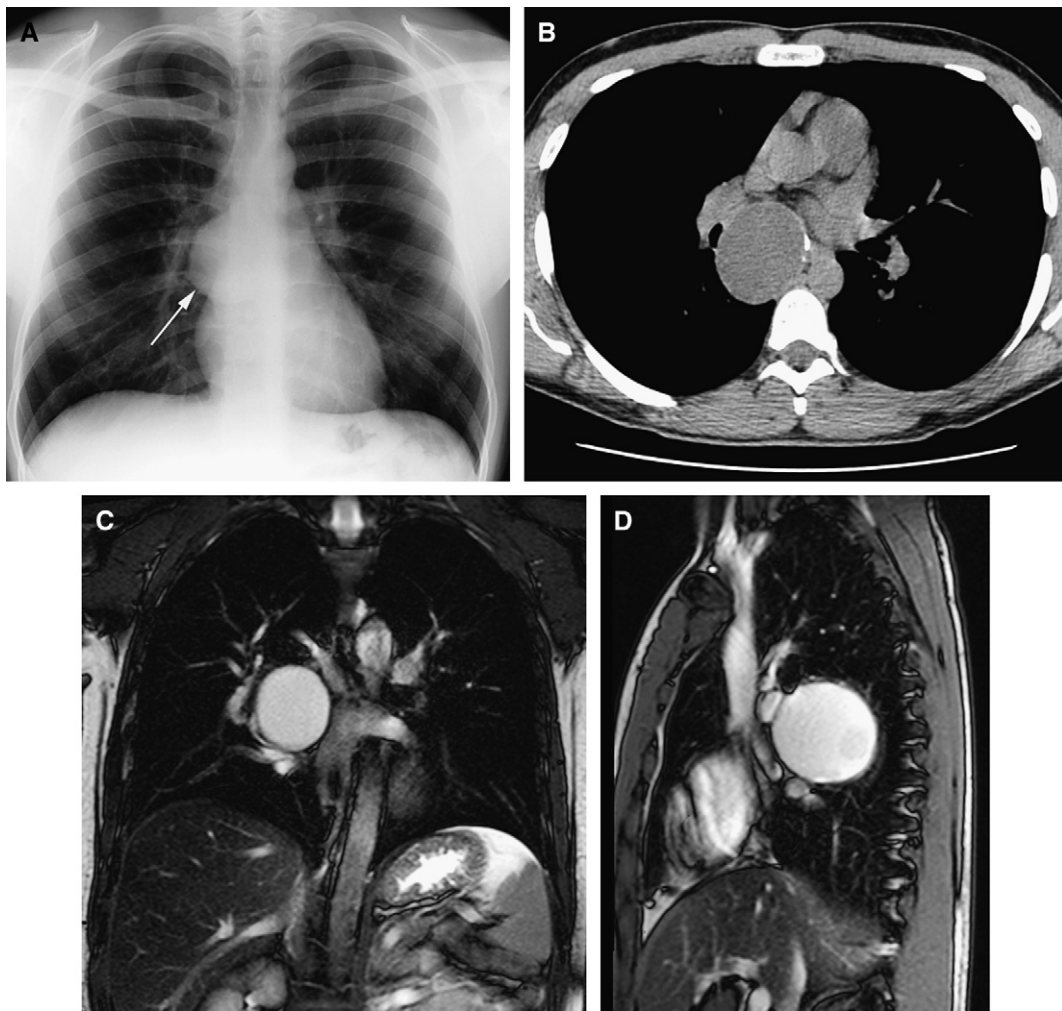


Figure 5. A 43-year-old woman who presented with dysphagia. (A) Chest radiograph, showing a smoothly margined soft tissue density mass beneath the carina (arrow). (B) Transaxial non-contrast-enhanced CT, showing a well-defined soft-tissue attenuation lesion. (C) Coronal and (D) sagittal T2-weighted MRI images, showing a thin-walled, uniformly high-signal lesion in the middle mediastinum.

Bronchogenic Cyst

Bronchogenic cysts arise from abnormal budding of the ventral foregut during the first trimester. They are thin walled, fluid filled, and lined by ciliated columnar epithelium [11]. Approximately two thirds of bronchogenic cysts are sited within the mediastinum, most often in a subcarinal or right paratracheal location. The remainder are intrapulmonary, with a predilection for the lower lobes. On plain radiographs, they appear as a rounded mass, with a smooth or lobulated border (Figure 5A). CT further defines their position and anatomical relationships, and most display homogeneous water or soft-tissue attenuation (Figure 5B). Higher attenuation values may be seen secondary to thick proteinaceous mucus, intracystic hemorrhage, or calcium oxalate crystal deposition. A thin rim of mural enhancement may normally be observed, but the presence of a thick or irregular wall is not a typical feature and suggests an alternative diagnosis [12]. Bronchogenic cysts display variable

signal intensity on T1-weighted magnetic resonance imaging (MRI) sequences, depending upon cyst contents and are of uniformly high signal on T2-weighted imaging (Figure 5C and D).

Intrapulmonary bronchogenic cysts may develop a fistulous connection with the tracheobronchial tree and become secondarily infected. The development of air-fluid levels can mimic cavitary lesions, such as bronchogenic carcinoma [13]. There are no specific imaging findings that allow easy differentiation, but a review of the clinical history and previous imaging is often helpful. Mediastinal bronchogenic cysts rarely become infected but can produce symptoms, such as stridor, dysphagia, and intractable cough, as a result of local compressive effects. There are reports of subcarinal bronchogenic cysts, inducing myocardial ischemia by compression of the left main stem coronary artery [14]. Complete excision of adult bronchogenic cysts is recommended in most instances to confirm the diagnosis, relieve symptoms, and prevent complications.

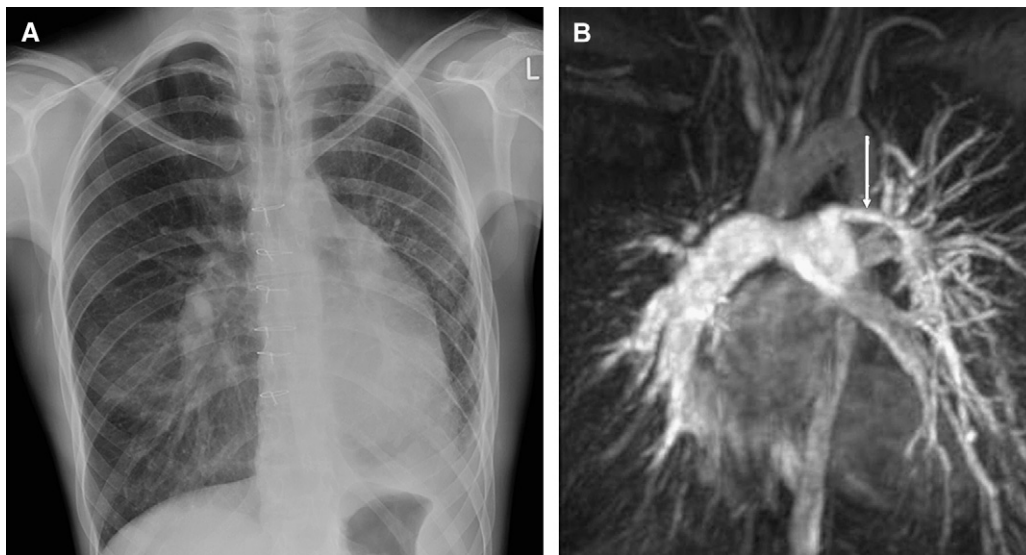


Figure 6. A 23-year-old woman with a history of left-sided congenital diaphragmatic hernia repair in early childhood. (A) Chest radiograph, showing a small left lung and leftward shift of the mediastinum. (B) MR pulmonary angiogram, showing a hypoplastic left pulmonary artery (arrow).

Abnormalities of the Lung Parenchyma

Pulmonary Agenesis/Aplasia

Pulmonary agenesis is the complete absence of a lung with no identifiable bronchus, parenchyma, or ipsilateral pulmonary artery. Aplasia is similar to agenesis, but there remains a rudimentary bronchus that ends in a blind pouch. Both conditions are rare and thought to be caused by interruption of blood flow in the dorsal aortic arch during early embryogenesis [15]. In the absence of associated congenital malformations in other organs, these conditions may be an incidental finding in adulthood. On plain radiographs, they simulate a pneumonectomy, with a small ipsilateral hemithorax, almost complete absence of aerated lung (depending on the degree of

compensatory hypertrophy), elevated hemidiaphragm, and displacement of the mediastinum to the affected side. CT confirms the absence of the ipsilateral pulmonary artery and can distinguish aplasia from agenesis by demonstrating a blind ending rudimentary bronchus [16].

Pulmonary Hypoplasia

Pulmonary hypoplasia represents deficient or incomplete development of the lung and is caused by factors that directly or indirectly compromise the thoracic space, such as congenital diaphragmatic hernia. Bronchi and alveoli are still present, but their growth is restricted. Most cases are detected at birth and survival largely depends upon the severity of any associated lesions [16].

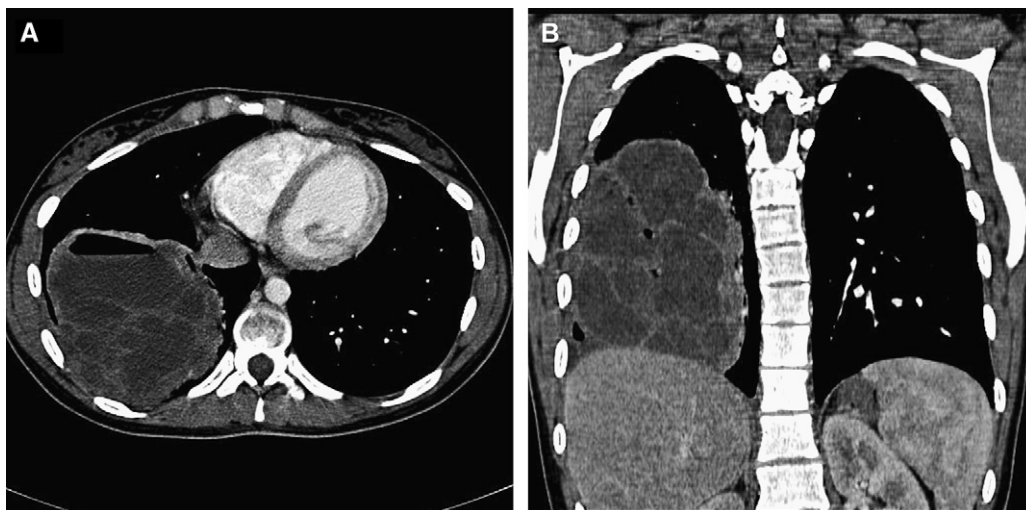


Figure 7. A 34-year-old woman who presented with fever and purulent sputum production. (A) Transaxial contrast-enhanced CT, showing a thin-walled multiloculated cystic lesion in the right hemithorax, which contains an air-fluid level. (B) Coronal reformatted image, showing the same. This lesion was surgically resected and histology confirmed a diagnosis of CAM.

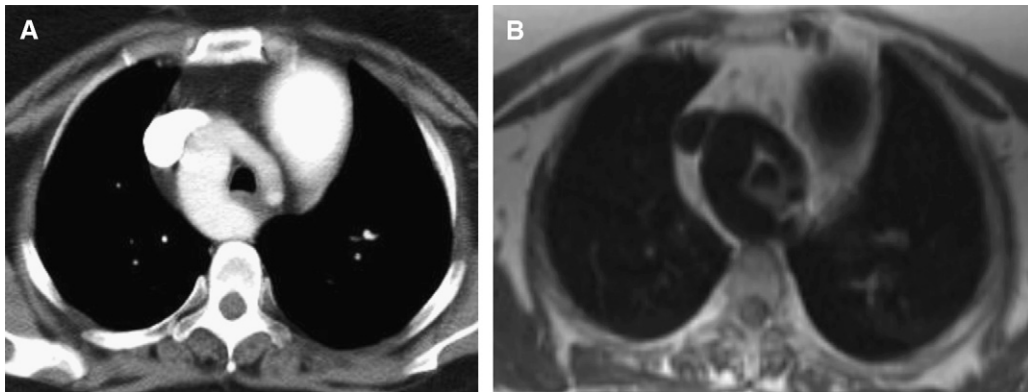


Figure 8. A 35-year-old woman who presented with dysphagia. (A) Transaxial contrast enhanced CT image, showing a vascular ring encircling the trachea and esophagus. (B) Transaxial T1-weighted “black blood” MRI sequence also showing the double aortic arch.

Plain radiographs demonstrate decreased aeration of the affected hemithorax, a small thoracic cage, and ipsilateral displacement of the mediastinum (Figure 6A) [16]. CT and MRI can be used to confirm the diagnosis and assess the degree of pulmonary underdevelopment. The ipsilateral pulmonary artery is present but of reduced calibre (Figure 6B) [17].

Cystic Adenomatoid Malformation

Cystic adenomatoid malformation (CAM) is an uncommon embryonic developmental abnormality that is usually diagnosed in infancy. On histologic examination, it is a malformation of the noncartilage containing terminal

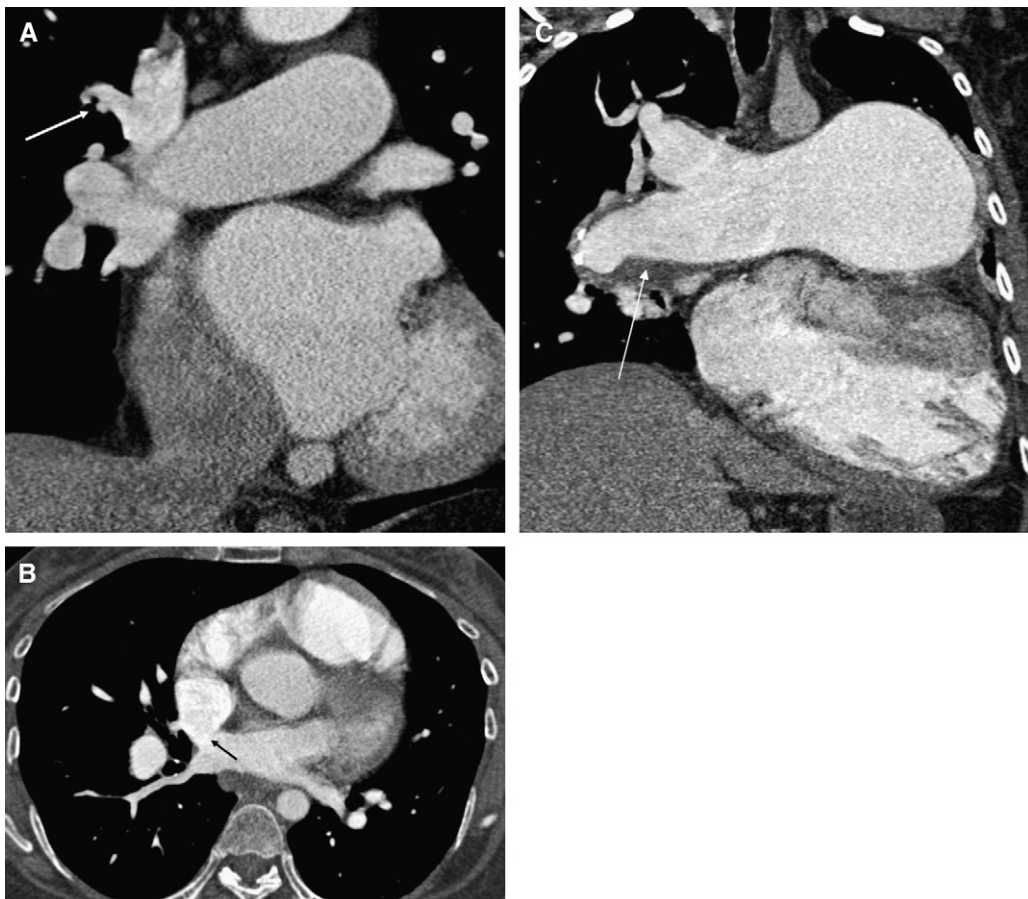


Figure 9. A 45-year-old woman with pulmonary hypertension. (A) Coronal reformatted CT image, showing partial anomalous pulmonary venous drainage of the right superior pulmonary vein into the superior vena cava (arrow). (B) Transaxial CT image, demonstrating an associated superior sinus venosus type atrial septal defect (arrow). (C) Coronal reformatted CT image, showing enlargement of the pulmonary artery, which contains partially calcified in situ thrombus (arrow).

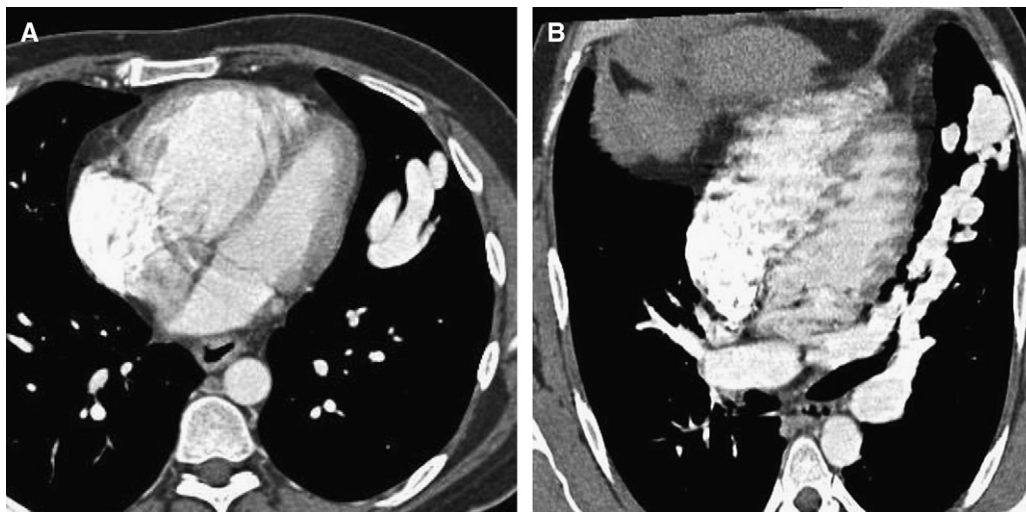


Figure 10. A 43-year-old man with hereditary hemorrhagic telangiectasia who presented after a transient ischemic attack. (A) Transaxial contrast-enhanced CT image, showing a large left-sided PAVM. (B) Maximum intensity projection reformat, showing an enhancing tortuous lesion, which connects with the left hilar vessels.

respiratory structures that communicates with the tracheo-bronchial tree and is supplied by a direct branch of the pulmonary artery [18]. Presentation in adulthood is rare, but it can cause recurrent chest infections, hemoptysis, or, more rarely, with malignant transformation in incompletely resected lesions of childhood. Sarcomas and bronchoalveolar carcinoma have been reported in association with CAM [19,20]. Chest radiographs demonstrate a cystic parenchymal lesion, and air-fluid levels may be seen if secondarily infected. CT confirms the presence of a thin-walled cystic lesion, which can appear more complex or even solid if secondarily infected (Figure 7) [21]. An absence of air bronchograms helps differentiate infected CAM from necrotizing pneumonia [15].

Abnormalities of the Vasculature

Vascular Rings

Tracheal compression can occur as a result of anomalous development of the aorta and its branches [15]. Double aortic arch is the most common type of complete vascular ring and consists of anterior and posterior arches that encircle the trachea and esophagus before joining distally to form a common descending aorta. This anomaly most commonly presents in childhood with stridor and dysphagia, but presentation in adulthood is also recognized. CT or MR angiography are the modalities of choice for demonstrating the vascular ring and degree of tracheal compression (Figure 8).

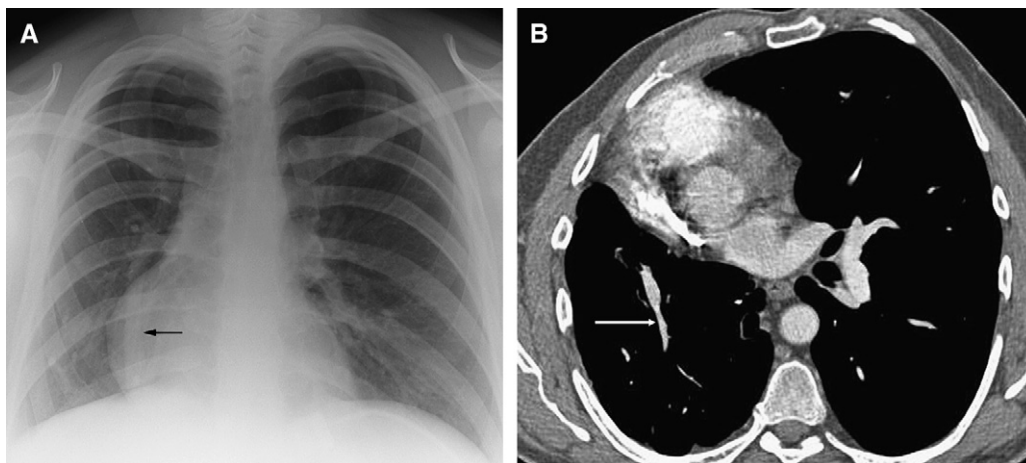


Figure 11. An asymptomatic 34-year-old woman. (A) Chest radiograph, showing loss of volume in the right hemithorax, rightward shift of the mediastinum, and a vessel is seen coursing inferiorly from the right hilum towards the diaphragm (arrow). (B) Transaxial contrast-enhanced CT image, showing a hypoplastic right lung and the anomalous vein coursing through it (arrow).

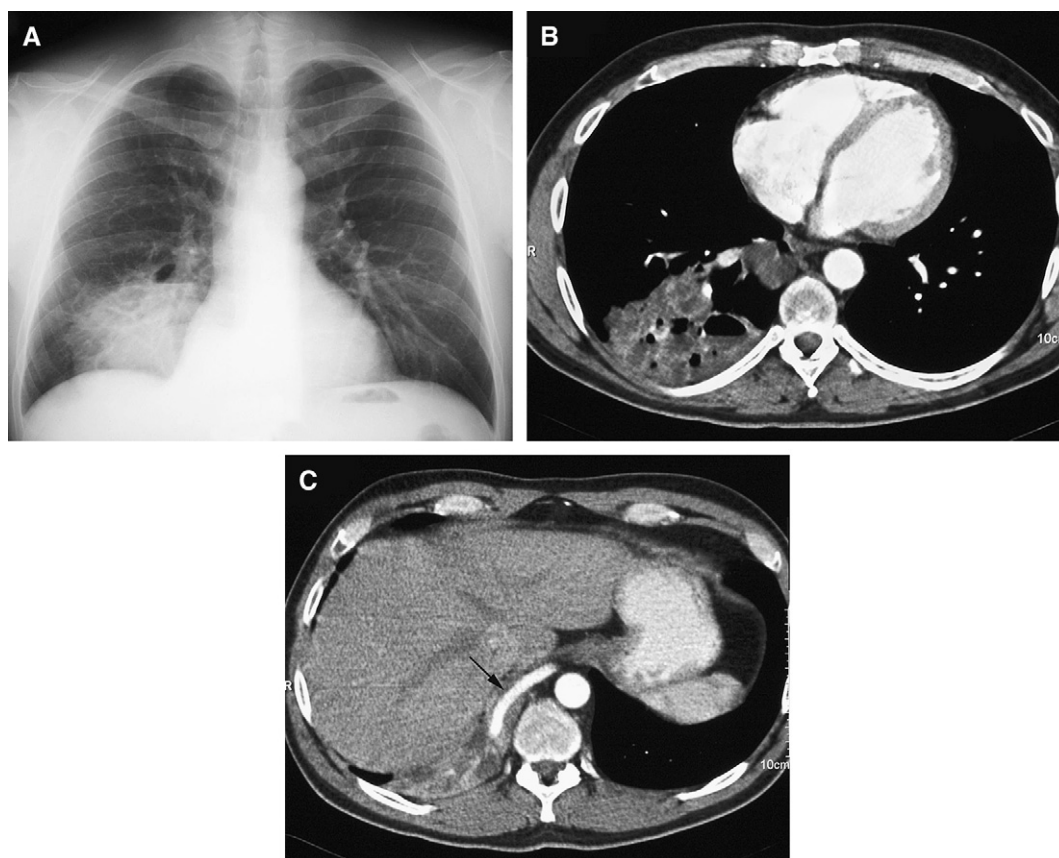


Figure 12. A 21-year-old man with recurrent chest infections. (A) Chest radiograph, showing a mass that contained multiple air-fluid levels in the right lower zone. (B) Transaxial contrast-enhanced CT image confirms the presence of a cystic-solid mass in the right lower lobe. (C) Transaxial CT image at a more caudal level, showing an anomalous vessel arising from the thoracic aorta, which supplies the sequestration (arrow).

Partial Anomalous Pulmonary Venous Return

Unlike total anomalous pulmonary venous return, partial anomalous pulmonary venous return (PAPVR) is an acyanotic lesion. It has a prevalence of 0.2% [22] and most commonly involves aberrant drainage of the right superior pulmonary vein into the superior vena cava to cause an extracardiac left-to-right shunt [8]. There is a frequent association with superior sinus venous type atrial septal defect, and patients may present with pulmonary hypertension as a consequence of these combined lesions. CT angiography readily demonstrates the anomalous venous drainage and may also detect any associated septal defect (Figure 9).

Pulmonary Arteriovenous Malformation

Pulmonary arteriovenous malformation (PAVM) can occur in isolation or as part of a systemic process in hereditary hemorrhagic telangiectasia [23]. They provide a direct communication between a pulmonary artery and vein, and the absence of a filtering capillary bed offers the potential for paradoxical embolism. They tend to be recognized with increasing age, because time is required for the effects of the shunt to become manifest [24]. Stroke may be the presenting feature, and percutaneous embolization is the treatment of

choice for avoidance of neurologic complications. CT angiography has a high sensitivity for diagnosing arteriovenous malformations and plays an important role in treatment planning [25]. PAVMs appear as an enhancing nodule or serpiginous mass connected with blood vessels (Figure 10) [26].

Combined Parenchymal/Vascular Abnormalities

Scimitar Syndrome

Scimitar syndrome encompasses a variety of congenital abnormalities of the thorax that occur in combination. Major components include a hypoplastic lung and pulmonary artery, partial systemic arterial supply via the aorta or its main branches, and partial anomalous pulmonary venous drainage [27]. It almost exclusively involves the right lung, and the draining vein most often empties into the infra-diaphragmatic inferior vena cava, although drainage to the hepatic veins, portal vein, and right atrium have been described [8]. The left-to-right shunt is usually hemodynamically insignificant, and most patients with Scimitar syndrome are asymptomatic and have a normal life span [28].

Chest radiographs show a small right hemithorax, rightward shift of the heart, and mediastinum and a curvilinear tubular opacity coursing inferiorly from the right hilum

towards the diaphragm (Figure 11A). This anomalous vein has been likened to the Scimitar, a curved blade used by Persian and Turkish warriors [28]. Contrast-enhanced CT or MRI can also be used to demonstrate the anomalous vessel and its anatomical connection (Figure 11B) [29].

Pulmonary Sequestration

Pulmonary sequestration is a mass of nonfunctioning lung tissue that is not in normal continuity with the tracheo-bronchial tree and receives a systemic arterial supply [30]. It arises when a supernumerary lung bud develops in early fetal life and the systemic artery is thought to be a persistent primitive aortic branch [31]. Sequestrations almost always occur in the lower lobes and are slightly more common in the left lung. They are divided into extralobar and intralobar types, depending on their pleural investment and pattern of venous drainage. Extralobar sequestrations are rare and consist of a discrete accessory lobe of nonaerated lung tissue that is invested in its own pleural envelope [30]. Most are located immediately above or below the left hemidiaphragm and have systemic venous drainage via the inferior vena cava or azygous system. They are usually diagnosed in early childhood as a consequence of the shunt and other associated congenital abnormalities [32]. Intralobar sequestrations are more common and consist of a segment of nonfunctioning lung tissue that is enclosed by the visceral pleura of an otherwise normal lung. Systemic arterial supply is usually via a single large artery arising from the lower thoracic or upper abdominal aorta [25]; however, arterial supply from other arteries, including the coronaries, has been described [33]. Venous drainage is via pulmonary veins into the left atrium. Intralobar sequestrations typically present in adulthood with recurrent pneumonias in a persistent location because of insufficient drainage. Bleeding within the sequestered segment is also common as a consequence of high pressure within the feeding artery; hemothorax and life-threatening torrential hemoptysis have been reported [34]. Sequestrations can mimic a range of thoracic pathology, including bronchogenic carcinoma, lung abscess, and neurogenic tumours. Plain radiographs reveal a lower lobe soft tissue density mass, which often contains air fluid levels, especially if infected (Figure 12A). CT angiography shows enhancement of the mass and in most cases demonstrates the anomalous feeding artery supplying it (Figure 12B and C). Multiplanar reformats are useful to show the pattern of venous drainage, which helps surgical planning by distinguishing extralobar from intralobar types [35].

Conclusion

A number of congenital thoracic lesions can present for the first time in adulthood and should be considered in the differential diagnosis of refractory pneumonias and mass lesions. Awareness of their typical imaging features will enable a confident diagnosis and help direct appropriate patient management.

References

- [1] Sasaki H, Yano M, Kaji M, et al. Anomalous origin of right upper lobe bronchus in a patient with lung cancer. *J Bronchology* 2003;4:289–91.
- [2] Ghaye B, Szapiro D, Fanchamps JM, et al. Congenital bronchial abnormalities revisited. *RadioGraphics* 2001;21:105–19.
- [3] McLaughlin FJ, Strieder DJ, Harris GB, et al. Tracheal bronchus: association with respiratory morbidity in childhood. *J Pediatr* 1985;106:751–5.
- [4] Gay S, Dee P. Tracheobronchomegaly: the Mounier-Kuhn syndrome. *Br J Radiol* 1984;57:640–4.
- [5] Johnston RF, Breen Tracheobronchomegaly RA. Report of five cases and demonstration of familial occurrence. *Am Rev Respir Dis* 1964;91:35–50.
- [6] Lazzarini-de-Oliveira LC, Costa de Barros Franco CA, Gomes de Salles CL, et al. A 38-year-old man with tracheomegaly, tracheal diverticulosis, and bronchiectasis. *Chest* 2001;120:1018–20.
- [7] Petrozzi M, Gilkeson R, McAdams HP, et al. Bronchial atresia: clinical observations and review of the literature. *Clin Pulm Med* 2001;8:101–7.
- [8] Zylak CJ, Eyer WR, Spizarny DL, et al. Developmental lung anomalies in the adult: radiologic-pathologic correlation. *RadioGraphics* 2002;22:25–43.
- [9] Tsuchida M, Aoki K, Hashimoto T, et al. Segmental bronchial atresia of the left upper lobe treated with segmental resection under video-assisted thoracic surgery. *Surg Laparosc Endosc Percutan Tech* 2001;11:217–20.
- [10] Kawamoto S, Yuasa M, Tsukuda S, et al. Bronchial atresia: three-dimensional CT bronchography using volume rendering technique. *Radiat Med* 2001;19:107–10.
- [11] Nuchtern JG, Harberg FJ. Congenital lung cysts. *Semin Pediatr Surg* 1994;3:223–43.
- [12] Ashizawa K, Okimoto T, Shirafuyi T, et al. Anterior mediastinal bronchogenic cysts: demonstration of complicating malignancy by CT and MRI. *Br J Radiol* 2001;74:959–61.
- [13] Aktogu S, Yuncu G, Halilcilar H. Bronchogenic cysts: clinicopathological presentation and treatment. *Eur Respir J* 1996;9:2017–21.
- [14] Kennebeck GA, Wong AK, Berry WR. Mediastinal bronchogenic cyst manifesting as a catastrophic myocardial infarction. *Ann Thorac Surg* 1999;67:1789–91.
- [15] Agrons GA, Courtney SE, Stocker JT, et al. From the archives of the AFIP: lung disease in premature neonates: radiologic-pathologic correlation. *RadioGraphics* 2005;25:1047–73.
- [16] Page DV, Stocker JT. Anomalies associated with pulmonary hypoplasia. *Am Rev Respir Dis* 1982;125:216–21.
- [17] Mutlu H, Basekim C, Silit E, et al. Gadolinium-enhanced 3D MR angiography of pulmonary hypoplasia and aplasia. *AJR Am J Roentgenol* 2006;187:398–403.
- [18] Zach MS, Eber E. Paediatric origins of adult lung lesions: adult outcome of congenital lower respiratory tract malformations. *Thorax* 2001;56:65–72.
- [19] Ueda K, Gruppo R, Unger F, et al. Rhabdomyosarcoma of lung arising in a congenital cystic adenomatoid malformation. *Cancer* 1977;40:383–8.
- [20] Ribet ME, Copin MC, Soots JG, et al. Bronchioloalveolar carcinoma and congenital cystic adenomatoid malformation. *Ann Thorac Surg* 1995;60:1126–8.
- [21] Shady K, Siegel MJ, Glazer HS. CT of focal pulmonary masses in childhood. *RadioGraphics* 1992;12:505–14.
- [22] Haramati LB, Moche IE, Rivera VT, et al. Computed tomography of partial anomalous pulmonary venous connection in adults. *J Comput Assist Tomogr* 2003;27:743–9.
- [23] Pelage JP, El Hajjam M, Lagrange C, et al. Pulmonary artery interventions: an overview. *RadioGraphics* 2005;25:1653–67.
- [24] Swanson KL, Udaya B, Prakash S, et al. Pulmonary arteriovenous fistulas: Mayo Clinic experience 1982–1997. *Mayo Clin Proc* 1999;74:671–80.
- [25] Engelke C, Schaefer-Prokop C, Schirg E, et al. High-resolution CT and CT angiography of peripheral pulmonary vascular disorders. *RadioGraphics* 2002;22:739–64.
- [26] Remy J, Remy-Jardin M, Giraud F, et al. Angioarchitecture of pulmonary arteriovenous malformations: clinical utility of three-dimensional helical CT. *Radiology* 1994;191:657–64.
- [27] Woodring JH, Howard TA, Kanga JF. Congenital pulmonary venolobar syndrome revisited. *RadioGraphics* 1994;14:349–69.

- [28] Ferguson EC, Krishnamurthy R, Oldham SAA. Classic imaging signs of congenital cardiovascular abnormalities. *Radiographics* 2007;27:1323–34.
- [29] Remy-Jardin M, Remy J. Spiral CT angiography of the pulmonary circulation. *Radiology* 1999;212:615–36.
- [30] Rosado-de-Christenson ML, Frazier AA, Stocker JT, et al. From the archives of the AFIP. Extralobar sequestration: radiologic-pathologic correlation. *RadioGraphics* 1993;13:425–41.
- [31] Gerle RD, Jaretzki A, Ashley CA, et al. Congenital bronchopulmonary-foregut malformation: pulmonary sequestration communicating with the gastrointestinal tract. *N Engl J Med* 1968;278:1413–9.
- [32] Nuchtern JG, Harberg FJ. Congenital lung cysts. *Semin Pediatr Surg* 1994;3:223–43.
- [33] Nakayama Y, Kido M, Minami K, et al. Pulmonary sequestration with myocardial ischemia caused by vasospasm and steal. *Ann Thorac Surg* 2000;70:304–5.
- [34] Miller EJ, Singh SP, Cerfolio RJ, et al. Pryce's type I pulmonary intralobar sequestration presenting with massive hemoptysis. *Ann Diagn Pathol* 2001;5:91–5.
- [35] Lee EY, Siegel MJ, Sierra LM, et al. Evaluation of angioarchitecture of pulmonary sequestration in pediatric patients using 3D MDCT angiography. *AJR Am J Roentgenol* 2004;183:183–8.